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PATIENTS WITH TRANSTHYRETIN (TTR) AMYLOIDOSIS HAVE A HIGHER MYOCARDIAL EXTRACELLULAR VOLUME (ECV) FRACTION THAN AL AMYLOID AS ASSESSED BY CARDIAC MAGNETIC RESONANCE (CMR) IMAGING: A PRELIMINARY INVESTIGATION

Poster Contributions
Hall C
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Authors: *Ashish Aneja, Emily Ruden, Pradeep Bhat, Subha Raman, MetroHealth Medical Center, Case Western Reserve University, Cleveland, OH, USA, The Ohio State University Medical Center, Columbus, OH, USA*

Background: Cardiac Amyloidosis (CA) is uncommon but has high morbidity and mortality. Biopsy remains the cornerstone of management. CMR is important in diagnosing CA but its ability to distinguish between CA subtypes has not been studied.

Methods: Patients diagnosed with amyloidosis from 1/2005 to present were selected from a hospital database. After cross-referencing with the CMR database, 65 patients had confirmed CA. Charts were reviewed for demographic, biopsy and vital status.

Results: Of the 65 patients with abnormal CMR and suspected CA, 33 had biopsy proven AL amyloid (lambda 22; kappa 11; 2 unknown), 16 had TTR amyloid (Val122Ile, 6; wild type 7; unknown 2; thr60ala 1), 1 Finnish type, 1 AA amyloid, 2 unknown; 44 men, 21 women; 49 White, 15 African-American, 1 of unknown race. Raw CMR images were reviewed and analyzed – 16 patients had both pre- and post-contrast T1 maps and pre-contrast T2 maps available, 3 had pre-contrast T1 and T2, 4 pre- and post-contrast T1, and 11 pre-contrast T2. Analysis revealed that TTR patients had significantly higher myocardial ECV (Table 1). An ROC curve model discriminated the two CA subtypes with an AUC of 0.864 +/-0.087 using 48% as the optimal ECV cut point.

Table 1.

	AL	TTR	P
ECV	40.1±1.2	58.0±1.1	<0.05
T1Blood pre	1508±228	1559±149	0.97
T1blood post	326±82	286±82	0.27
T1myo pre	1000±151	1078±65	0.13
T1myopost	387±93	286±83	0.04
T2Blood	230±64	239±67	0.86
T2Myo	62±7	61±10	0.39
Hematocrit	0.36 +/- 0.06	0.36 +/- 0.06	0.72

Conclusion: Patients with TTR CA have significantly higher ECV than patients with AL CA. This phenomenon may result from the indolent course of TTR CA, leading to delayed clinical presentation and greater cardiac involvement at the time of CMR study. These important findings need confirmation in a prospective study.